CASE REPORT
DOI 10.4070/kcj.2011.41.6.331

Print ISSN 1738-5520 / On-line ISSN 1738-5555

Open Access

Surgical Repair of Giant Right Atrial Aneurysm in a Neonate

Youngok Lee, MD[1], Joonyong Cho, MD[1], Gunjik Kim, MD[1],
Sangbum Lee, MD[2], Myungchul Hyun, MD[2], and Yeohyang Kim, MD[3]

[1]Departments of Thoracic and Cardiovascular Surgery and [2]Pediatric Cardiology, Kyungpook National University College of Medicine, Daegu, [3]Department of Pediatric Cardiology, Keimyung University College of Medicine, Dongsan Hospital, Daegu, Korea

ABSTRACT

Right atrial aneurysm is an extremity rare congenital heart defect. We report a case which was diagnosed during the early fetal stage and operated on in a 2 week old neonate. Following median sternotomy, aneurysmectomy was performed under a cardiopulmonary bypass. Histopathological examination of the resected atiral tissue showed a lipotomatous degeneration and reduction of the muscular elements. There were no postoperative complications. **(Korean Circ J 2011;41:331-333)**

KEY WORDS: Right atrium; Heart aneurysm.

Introduction

Isolated aneurysm of the right atrium (RA) is a rare congenital abnormality and its aneurismal resection was first described by Morrow and Behrendt.[1)] Since then, a few cases of RA aneurysmectomy have been reported in the literature. Besides one case, no patients underwent an operation in the neonatal period.[2)] We report a case which was diagnosed during the early fetal stage and operated on in a 2 week old neonate with excision of the aneurysm.

Case

A 2 week old neonate, 3.5 kg in weight, was referred to the hospital for a giant aneurysm on the RA discovered during a routine prenatal ultrasound. There was no fetal distress and the delivery was vaginal at term. The patient was asymptomatic and the physical examination was entirely normal. The electrocardiography showed a normal sinus rhythm and the chest X-ray revealed a cardiothoracic ratio of 80% with normal pulmonary vascular marking (Fig. 1). Transthoracic echocardiography revealed a giant right atrial aneurysm which measured 5.4×3.6 cm^2 and occupied the lateral and anterior wall of the RA, extending in to the anterior wall of the right ventricle (Fig. 2). The right ventricle was compressed but its inflow was not impaired and the tricuspid valve was not displaced. No intracavitary thrombus was indentified. The remainder of the study was normal.

Since the risk of surgical resection is low and the natural clinical course of a right atrial aneurysm remains unclear, we decided to do surgical management before the emergence of any complications, such as arrhythmia or a thromboembolic event.

Standard median sternotomy was performed. The pericardial sac was complete and opening of the pericardium revealed a giant aneurysm occupying the free wall of the RA which measured approximately 5×6 cm^2 (Fig. 3). The wall of the aneurysm was thin, floppy and non-contractile, and the right ventricle was significantly compressed by the aneurysm.

After establishing a standard cardiopulmonary bypass, the aneurysm was opened externally. Thrombus or any inflammatory signs were not seen in the aneurysm or in the RA, and no other cardiac abnormalities except patent foramen ovale were found. After excision of the affected right atrial wall, the defect was over sewn from the RA.

The patent foramen ovale was also closed by direct suture. Histopathological examination of the resected atrial tissue showed a lipotomatous degeneration and reduction of the muscular elements and no evidence of inflammatory reaction.

Received: July 31, 2010
Revision Received: September 28, 2010
Accepted: October 20, 2010
Correspondence: Joonyong Cho, MD, Department of Thoracic and Cardiovascular Surgery, Kyungpook National University College of Medicine, 200 Dongdeok-ro, Jung-gu, Daegu 700-721, Korea
Tel: 82-53-420-5665, Fax: 82-53-426-4765
E-mail: jycho@knu.ac.kr

• The authors have no financial conflicts of interest.

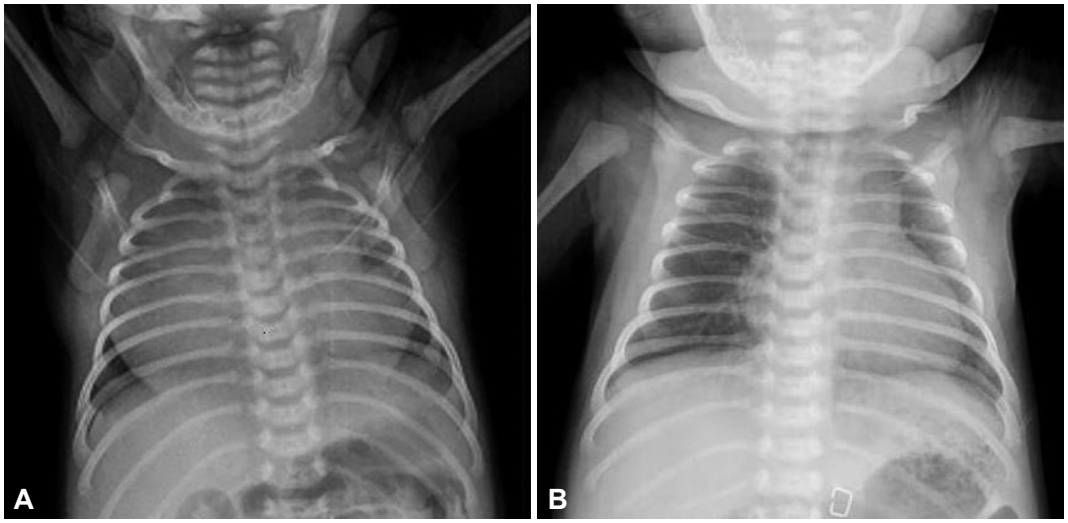

Fig. 1. Preoperative (A) and postoperative X-rays (B) demonstrating a significant reduction in cardiac size.

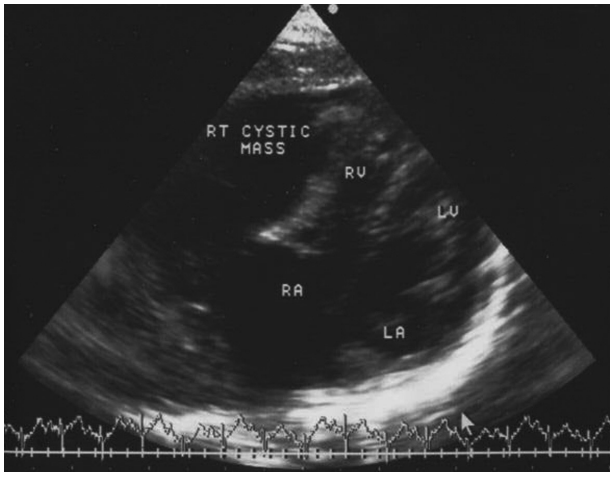

Fig. 2. Preoperative echocardiography demonstrating a giant right atrial aneurysm.

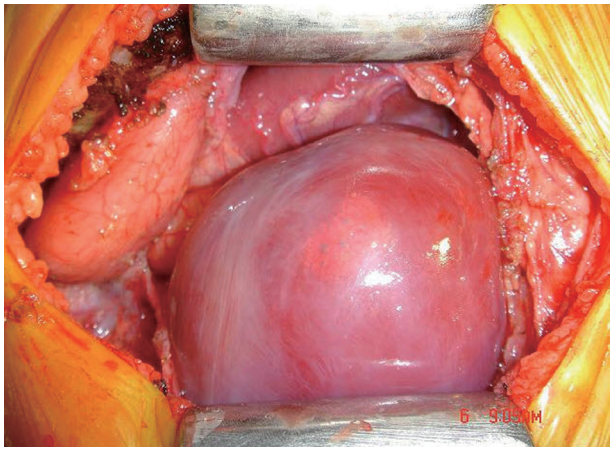

Fig. 3. Intraoperative view showing the massively dilated right atrium.

The post-operative course was uneventful and the post-operative X-ray showed a significant reduction in cardiac size (Fig. 1). The patient was in good health during 2 years of a post-operative follow up course and the echocardiography sowed normal contractility and size of the RA.

Discussion

An atrial aneurysm is defined as the dilatation of the atrium involving all layers of the atrial wall. Aneurysms of the RA are very rare and may be congenital or acquired. In the absence of predisposing conditions such as tricuspid valve disease, congenital heart disease, pulmonary arterial hypertension or acquired inflammatory changes in the myocardium, aneurysms of the RA are considered to be of congenital origin.[3] The origin of a congenital aneurysm is unknown. In some cases, it has been described as a congenital structural defect in the atrial wall that makes it prone to dilatations even with right sided pressure that is lower than the left side. It was reported that an intrinsic factor may play a role as tissue weakening due to a protein defect or as deficiency of collagen type III (e.g., Ehlers-Danlos syndrome type IV).[3][4] Others have attributed it to dysplasia of the pectinate muscles.[5]

The age at presentation varies from the neonatal period to late adulthood. Many patients are asymptomatic. The most common symptoms are palpitations due to atrial tachycardia, including atrial flutter and fibrillation. Other rhythm abnormalities include pre-excitation, junctional rhythm, atrioventricular block and incessant supraventricular tachycardia.[3] In addition, potential complications include pulmonary thrombo-embolism, paradoxical systemic embolization and atrial rupture.[2-7] It was reported that 5 deaths occurred in patients with right atrial aneurysm.[3]

The diagnosis of right atrial aneurysm can be established with echocardiography, angiography, computer tomography (CT) or magnetic resonance imaging (MRI). Transthoracic echocardiography is the most commonly used technique. At times, it may be difficult to distinguish atrial aneurysm from contiguous tumors of the heart. CT and MRI of the chest may

be helpful for a definitive diagnosis.[3)6)]

The optimal therapeutic approach for right atrial aneurysms is still under debate. Surgical resection, catheter-based ablation, and follow up with conservative treatment are some therapeutic options. If patients do not receive surgical resection, anti-coagulation is crucial because of the increased risk of thrombus development in the right atrial aneurysm.

Due to the potential risk of life threatening complications (pulmonary embolism, atrial rupture, sudden death), we strongly recommend early surgical resection before significant atrial anatomic and electrophysiological remodeling occurs. The resection may be carried out with or without cardiopulmonary bypass.[2-7)] However, authentic resection with the support of cardiopulmonary support is a safer method, especially in pediatric patients. We believe that very low operative mortality rates and high success rates justify a surgical approach in patients with atrial aneurysm. Also, surgical resection will provide an active life without anti-coagulation treatments.

REFERENCES

1) Morrow AG, Behrendt DM. *Congenital aneurysm (diverticulum) of the right atrium: clinical manifestations and results of operative treatment. Circulation 1968;38:124-8.*
2) Mascio CE, Burkhart HM, Fagan T, Ge S, Burkhart DM. *Neonatal right atrial aneurysm. Ann Thorac Surg 2005;80:2382.*
3) Binder TM, Rosenhek R, Frank H, Gwenchenberger M, Maurer G, Baumgartner H. *Congenital malformations of the right atrium and the coronary sinus: an analysis based on 103 cases reported in the literature and two additional cases. Chest 2000;117:1740-8.*
4) Zeebregts CJ, Hensens AG, Lacquet LK. *Asymptomatic right atrial aneurysm: fortuitous finding and resection. Eur J Cardiothorac Surg 1997;11:591-3.*
5) Papagiannis J, Chatzis A, Sarris G. *Giant right atrial aneurysm. Int J Cardiol 2008;129:e5-6.*
6) Moraes F, Santos CL, Lira V, Moraes CR. *Congenital aneurysm of the right atrium. Eur J Cardiothorac Surg 2001;19:943-4.*
7) Klisiewicz A, Szymanski P, Rozanski J, Hoffman P. *Giant congenital aneurysm of the right atrium: echocardiographic diagnosis and surgical management. J Am Soc Echocardiogr 2004;17:286-7.*